

## Complete Summary

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### GUIDELINE TITLE

Systemic lupus erythematosus (SLE).

### BIBLIOGRAPHIC SOURCE(S)

Gripenberg-Gahmberg M. Systemic lupus erythematosus (SLE). In: EBM Guidelines. Evidence-Based Medicine [CD-ROM]. Helsinki, Finland: Duodecim Medical Publications Ltd.; 2004 Aug 12 [various]. [11 references]

### GUIDELINE STATUS

This is the current release of the guideline.

This guideline updates a previous version: Finnish Medical Society Duodecim. Systemic lupus erythematosus (SLE). Helsinki, Finland: Duodecim Medical Publications Ltd.; 2001 Apr 30. Various p.

## COMPLETE SUMMARY CONTENT

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## SCOPE

### DISEASE/CONDITION(S)

Systemic lupus erythematosus (SLE)

### GUIDELINE CATEGORY

Diagnosis  
 Treatment

### CLINICAL SPECIALTY

Family Practice  
Internal Medicine  
Rheumatology

## INTENDED USERS

Health Care Providers  
Physicians

## GUIDELINE OBJECTIVE(S)

Evidence-Based Medicine Guidelines collects, summarizes, and updates the core clinical knowledge essential in general practice. The guidelines also describe the scientific evidence underlying the given recommendations.

## TARGET POPULATION

Patients with or suspected to have systemic lupus erythematosus (SLE)

## INTERVENTIONS AND PRACTICES CONSIDERED

### Diagnosis

1. Assessment of clinical features
2. Laboratory evaluation (blood count, platelets, sedimentation rate, anti-nuclear antibodies, dipstick test of the urine and urinalysis)
3. American Rheumatism Association (ARA) classification
4. Referral to a specialist, as indicated, for evaluation

### Treatment

1. Individualized treatment depending on the manifestations and activity of the disease
2. Patient education: avoidance of sunbathing, use of sunscreens
3. Pharmacologic therapy (nonsteroidal anti-inflammatory drugs, hydroxychloroquine, corticosteroids, immunosuppressive drugs [e.g. azathioprine, cyclophosphamide])
4. Other drugs as indicated, such as antihypertensive treatment
5. Treatment of discoid lupus with fluocinonide cream, hydrocortisone cream, hydroxychloroquine, or acitretin
6. Referral to nephrologist for signs of renal manifestations

## MAJOR OUTCOMES CONSIDERED

- Risk of relapse
- Risk for mortality
- Risk for end-stage renal disease
- Degree of clearing/improvement of discoid lupus
- Lupus signs and symptoms

## METHODOLOGY

### METHODS USED TO COLLECT/SELECT EVIDENCE

Hand-searches of Published Literature (Primary Sources)  
Hand-searches of Published Literature (Secondary Sources)  
Searches of Electronic Databases

### DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

The evidence reviewed was collected from the Cochrane database of systematic reviews and the Database of Abstracts of Reviews of Effectiveness (DARE). In addition, the Cochrane Library and medical journals were searched specifically for original publications.

### NUMBER OF SOURCE DOCUMENTS

Not stated

### METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

### RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence

- A. Strong research-based evidence. Multiple relevant, high-quality scientific studies with homogeneous results.
- B. Moderate research-based evidence. At least one relevant, high-quality study or multiple adequate studies.
- C. Limited research-based evidence. At least one adequate scientific study.
- D. No research-based evidence. Expert panel evaluation of other information.

### METHODS USED TO ANALYZE THE EVIDENCE

Review of Published Meta-Analyses  
Systematic Review

### DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

### METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

### RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

## COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

## METHOD OF GUIDELINE VALIDATION

Peer Review

## DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Not stated

# RECOMMENDATIONS

## MAJOR RECOMMENDATIONS

The levels of evidence [A-D] supporting the recommendations are defined at the end of the "Major Recommendations" field.

### Introduction

- Systemic lupus erythematosus (SLE) is a syndrome characterized by clinical diversity, changes in the disease activity over time, and by aberrant immunological findings.

### Epidemiology

- The prevalence of SLE worldwide is 4 to 250 per 100,000. The incidence is most frequent in women aged 15 to 25 years.

### Clinical Presentation

- The clinical presentation varies between different patients, and in a single patient the disease activity varies over time.
- Constitutional symptoms such as fatigue and fever are common.
- A vast majority of the patients have arthralgia, mostly of the hands.
- About one-half of the patients have cutaneous features, such as malar rash and discoid lupus as well as photosensitivity.
- About one-third of the patients have oral ulcerations.
- About 50% of the patients have nephropathy, which varies from mild proteinuria and microscopical hematuria to end-stage renal failure.
- About 20 to 40% of the patients have pleurisy. Acute pneumonitis and chronic fibrotising alveolitis are relatively rare.
- Pericarditis is somewhat more uncommon than pleuritis. T-wave changes in the electrocardiogram (ECG) are usual.
- Depression and headache are the most common of the neuropsychiatric symptoms. Grand-mal seizures and organic psychoses are rare. A peripheral

neuropathy is observed in about 10% of the patients and as many patients get a thromboembolic or hemorrhagic complication of the brain.

- The lymph nodes may enlarge especially when the disease is active.
- There is a risk of first and second trimester foetal losses and of premature birth.

### Laboratory Findings

- Laboratory findings are diverse.
- Sedimentation rate is usually elevated; the C-reactive protein (CRP) value is usually normal.
- Mild or moderate anaemia is common. A clear-cut haemolytic anaemia is seen in less than 10% of the patients.
- Leucocytopenia (lymphocytopenia)
- Mild thrombocytopenia
- Antinuclear antibodies are found in over 90% of the patients.
- Anti-deoxyribonucleic acid (DNA) antibodies (in 50–90% of the patients)
- Polyclonal hypergammablobulinaemia
- Decreased complement values (C3 and C4)
- Antiphospholipid antibodies
- Proteinuria, microscopic hematuria, decreased creatinine clearance

### Diagnosis

- There is no single symptom or finding that in itself is sufficient for making the diagnosis.
- When SLE is suspected the basic laboratory investigations are:
  - Blood count
  - Platelets
  - Sedimentation rate
  - Antinuclear antibodies
  - Dipstick test of the urine and urinalysis
- The diagnosis is based on the clinical symptoms and the laboratory findings and on the American Rheumatism Association (ARA) classification criteria (1982).
- The patient should be referred to a specialist for confirmation of the diagnosis.

### Treatment

- The treatment is always individual and depends on the manifestations and activity of the disease. There is no need for treatment solely on the basis of the immunological findings.
- The patients should be encouraged to restrain from sunbathing and to use sunscreens.
- The most important drugs are:
  - Nonsteroidal anti-inflammatory drugs
  - Hydroxychloroquine ("A randomized study of the effect," 1991; Wallace, 1994) [C]
  - Corticosteroids
  - Immunosuppressive drugs (e.g. azathioprine, cyclophosphamide)

- Hydroxychloroquine and nonsteroidal anti-inflammatory drugs are used in the treatment of mild symptoms such as cutaneous manifestations and arthralgia. When the response is insufficient or when the patient has fatigue or fever, a low dose of corticosteroids (prednisolone 5–7.5 mg/day) can be added.
- In the treatment of pleuritis or pericarditis, larger amounts of corticosteroids (about 30 mg prednisolone per day) are used.
- In the treatment of severe central nervous system (CNS) symptoms and of severe glomerulonephritis, thrombocytopenia, and hemolytic anaemia, large corticosteroid doses and other immunosuppressive drugs are used (Bansal & Beto, 1997; DARE, 1999; Flanc et al., 2004) [A].
- The differential diagnosis between an infection and a flare of the SLE is of utmost importance.
- Other drugs that the patient might need, such as antihypertensive treatment, should be remembered.
- If there are signs of renal manifestations, the patient should be referred to a nephrologist for a renal biopsy.
- The patients are often allergic to a variety of antibiotics, especially sulfonamides.

#### Primary Antiphospholipid Syndrome

- A syndrome manifesting as recurrent venous or arterial thrombotic events, recurrent miscarriages, thrombocytopenia, and antiphospholipid antibodies, but without other features of SLE

#### Related Evidence

- Fluocinonide cream is more effective than hydrocortisone for discoid lupus erythematosus (Jessop, Whitelaw, & Jordaan, 2002) [C]. Hydroxychloroquine and acitretin are as effective.

#### Definitions:

#### Levels of Evidence

- Strong research-based evidence. Multiple relevant, high-quality scientific studies with homogeneous results.
- Moderate research-based evidence. At least one relevant, high-quality study or multiple adequate studies.
- Limited research-based evidence. At least one adequate scientific study.
- No research-based evidence. Expert panel evaluation of other information.

#### CLINICAL ALGORITHM(S)

None provided

#### EVIDENCE SUPPORTING THE RECOMMENDATIONS

#### REFERENCES SUPPORTING THE RECOMMENDATIONS

[References open in a new window](#)

## TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

Concise summaries of scientific evidence attached to the individual guidelines are the unique feature of the Evidence-Based Medicine Guidelines. The evidence summaries allow the clinician to judge how well-founded the treatment recommendations are. The type of supporting evidence is identified and graded for select recommendations (see the "Major Recommendations" field).

## BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

### POTENTIAL BENEFITS

- Effective and safe treatment of systemic lupus erythematosus (SLE)
- Appropriate specialist referral

### POTENTIAL HARMS

Risk of Relapse

Discontinuing hydroxychloroquine medication in stable systemic lupus erythematosus (SLE) increases the risk of relapse.

## IMPLEMENTATION OF THE GUIDELINE

### DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

## INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

### IOM CARE NEED

Getting Better  
Living with Illness

### IOM DOMAIN

Effectiveness  
Patient-centeredness

## IDENTIFYING INFORMATION AND AVAILABILITY

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## ADAPTATION

Not applicable: The guideline was not adapted from another source.

## DATE RELEASED

2001 Apr 30 (revised 2004 Aug 12)

## GUIDELINE DEVELOPER(S)

Finnish Medical Society Duodecim - Professional Association

## SOURCE(S) OF FUNDING

Finnish Medical Society Duodecim

## GUIDELINE COMMITTEE

Editorial Team of EBM Guidelines

## COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Primary Author: Marianne Gripenberg-Gahmberg

## FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

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## GUIDELINE AVAILABILITY

This guideline is included in a CD-ROM titled "EBM Guidelines. Evidence-Based Medicine" available from Duodecim Medical Publications, Ltd, PO Box 713, 00101 Helsinki, Finland; e-mail: [info@ebm-guidelines.com](mailto:info@ebm-guidelines.com); Web site: [www.ebm-guidelines.com](http://www.ebm-guidelines.com).

## AVAILABILITY OF COMPANION DOCUMENTS

None available

## PATIENT RESOURCES



None available

## NGC STATUS

This summary was completed by ECRI on December 17, 2002. The information was verified by the guideline developer as of February 7, 2003. This NGC summary was updated by ECRI on October 4, 2004.

## COPYRIGHT STATEMENT

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